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Primary Antral Duodenal Extramedullary Plasmacytoma Presenting With Melena

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A 69-year old African woman presented with a 2-week history of fatigue and melena. Her medical history was notable for congestive heart failure complicated by an apical thrombus requiring Coumadin. Physical examination showed pallor, but was otherwise unremarkable. Abnormal laboratory values included a hemoglobin level of 4.4 g/dL and an international normalized ratio of 2.3.

An upper gastrointestinal endoscopy showed an erosive antral gastropathy (Figure A) and an ulcerated mass located on the posterior-superior wall of the duodenum (Figure B). Biopsy specimens from the antrum and duodenum showed a plasma cell infiltrate with plasmablastic features (Figure C, 400×, H&E). Neoplastic cells were reactive for the plasma cell marker CD138 and found by in situ hybridization to be lambda immunoglobulin light chain restricted (Figure C). CD20, a B-cell marker, was not expressed by the malignant cells. The morphology in conjunction with the immunohistochemistry was most consistent with an extramedullary plasmacytoma, and the negativity for the B-cell marker CD20 excluded the possibility of marginal zone B-cell lymphoma of mucosa associated lymphoid tissue lymphoma. A subsequent bone marrow biopsy showed a normo-cellular marrow with trilineage hematopoiesis with less than 5% plasma cells. Serum electrophoresis and immunofixation showed an M-spike of immunoglobulin A lambda, a finding identified in approximately 25% of plasmacytoma cases.¹ Together, these findings confirmed a final diagnosis of a primary extramedullary plasmacytoma of the duodenum and stomach as the cause of her melena.

Coumadin therapy was stopped, which limited her bleeding. Due to cardiac and renal comorbidities, the patient was deemed a poor candidate for surgical intervention or chemotherapy. Extra-corporeal radiation therapy was begun and initially reduced the overall size of her duodenal plasmacytoma. However, because of its location it ultimately progressed to a state of biliary obstruction. At endoscopic retrograde cholangiopancreatography, the bile duct was inaccessible secondary to tumor burden. The patient required a percutaneous transhepatic biliary drainage.

Extramedullary plasmacytoma is a rare entity accounting for less than 4% of all plasma cell tumors and occurs mainly in the upper aerodigestive tract. In the gastrointestinal tract, most tumors occur in the stomach, whereas only 20% to 30% arise in the small intestine.² No general consensus guidelines for treatment currently exist. However, based on the well-known radiation sensitivity of plasma cell tumors, radiotherapy is accepted as a standard therapy.³ Surgery for extramedullary plasmacytomas of the small intestine can also offer cure and may be the preferred treatment for some patients.⁴ If complete surgical resection is not feasible, then combined radiation or chemotherapy is recommended. The overall disease prognosis is good, with a 10-year overall survival of 70%.⁵ In a review of 721 cases by Alexiou et al,³ after treatment, approximately 65% of patients were free of recurrence and did not progress to multiple myeloma, 21% experienced recurrence, and 14% of cases evolved to multiple myeloma.

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